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# SOLITARY INTRACRANIAL ENCHONDROMA IN NON-SYNDROMIC SETTING

Pathology

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## ABSTRACT

Enchondromas are rare, benign cartilaginous neoplasm. Intracranial location is exceedingly rare. We present a solitary mass in intraaxial location in absence of Olliers or Mafucci's disease.

## KEYWORDS

enchondroma, intracranial, cerebral, non-syndromic

## INTRODUCTION

Intracranial enchondroma are extremely rare. Incidence is 13% in Maffucci's syndrome. The median age is 28 years (range 11–74 years) and 57.1% occur in female patients. Extracranial chondrosarcoma is reported in 3 (21.4%) patients. Common presenting symptoms of enchondroma include headache, seizure, and diplopia or detected incidentally. Common locations are petroclival fissure (86%) and clivus (71%). [1,2]

### Case Study

A 23-year-old male presented with seizures for 3 years. Radioimaging showed 6.4 x 5.6 x 5.4 cm well defined, lobulated mass in the right parieto-temporal lobe. It was predominantly cystic with peripheral walls showing nodular calcifications. Peripheral solid areas were isointense on T1 and hypointense on T2 with heterogenous contrast enhancement. Central cystic areas were hypointense on T1 and hyperintense on T2. (Figure 1a) Impression was old healed parasitic lesion or cystic meningioma. EN bloc resection was performed. A partially opened cytic mass with hard white pearly walls was received. (Figure 1b)

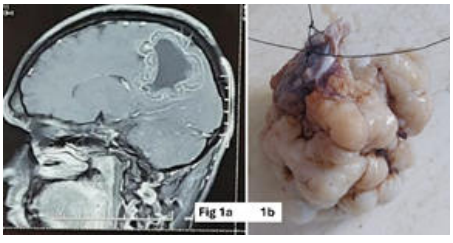


**Figure 1** a: Magnetic resonance imaging. Fig 1b shows gross appearance of tumor.

Histopathology showed encapsulated tumor with lobules of mature chondroid tissue. Cells lay within lacunae and embedded within chondroid matrix. There was absence of binucleate cells, cellular atypia, tumor necrosis or mitosis. There was absence of physaliferous cells. (Figure 2a,2b, 2c)

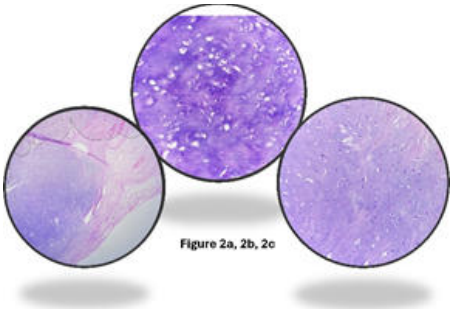


**Figure 2** a: Photomicrograph shows encapsulated mass with lobules of mature cartilage. 2b and 2c show cells in lacunae embedded in chondroid matrix.

Immunohistochemistry was positive for S100 but negative for pan cytokeratin and EMA. Ki67 index was less than 1 %. A final diagnosis of intracerebral enchondroma was offered. There was no recurrence or mortality upto 2 years of followup. There were no additional similar lesions in other bones, ruling out Olliers and Mafucci's disease.

## CONCLUSIONS

Enchondromas are benign hamartomatous neoplasms of chondroid tissue, usually as solitary, central, metaphyseal tumors of hands, feet and humerus. Skull base tumors are reported in petroclival region. Enchondroma tend to begin in childhood and become symptomatic by third decade. They tend to arise from chondrocyte cell plates with mutations in IDH1 or IDH2 which prevent the normal osteogenic differentiation of stem cells.

Ollier's disease has multiple enchondromas in asymmetric distribution in appendicular skeleton. Maffucci's syndrome has multiple enchondromas with soft tissue hemangioma and lymphangioma. Both syndromes are associated with sarcomatous transformation of enchondroma and visceral malignancies. IDH1 mutations make synchronous glioma existence a possibility. [1-4]

Our case is rare being intracranial, frontoparietal and in non-syndromic setting.

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